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The Role of Computed Tomography in the Diagnosis and Surveillance of Pepper Syndrome: A Case-Based Clinical Article

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Abstract **Case Report**

Background: Pepper syndrome is a clinical sub-entity of stage 4S neuroblastoma defined by primary adrenal neuroblastoma with massive, diffuse liver metastases in infants under 1 year of age. Computed Tomography (CT) plays a pivotal role in staging and therapeutic monitoring. ***Case Presentation:*** We report the case of a 10-month-old male infant presenting with general health alteration and mild abdominal distension. A thoraco-abdo-pelvic CT revealed a heterogeneous hypodense left adrenal mass measuring. The liver demonstrated multiple non-homogenous metastatic nodular. ***Conclusion:*** Multi-detector CT provides crucial objective measurement of target lesions, accurate anatomical staging, and assessment of vascular/organ compression, supporting decision-making in Pepper syndrome.

Keywords: Pepper Syndrome, Stage 4S Neuroblastoma, Adrenal Mass, Hepatic Metastases, Computed Tomography, Pediatric Radiology.

INTRODUCTION

Neuroblastoma is the most prevalent extracranial solid malignancy in pediatric oncology [1]. Stage 4S (or INRG Stage MS) represents a unique subgroup occurring in infants under 18 months, characterized by spontaneous regression potential despite extensive metastatic disease restricted to the liver, skin, or bone marrow [2, 3, 11]. Pepper syndrome specifically describes stage 4S neuroblastoma with overwhelming hepatic infiltration [4, 5]. CT imaging is critical for confirming the diagnosis, mapping target lesions, evaluating vascular integrity, and ruling out cortical bone involvement that would upstage the tumor to high-risk stage 4 [7, 8]. We present a detailed CT-based report of a 10-month-old infant to illustrate the clinical utility of CT in Pepper syndrome.

CASE PRESENTATION

A 10-month-old male infant presented due to persistent mild abdominal distension and a mild deterioration in general condition. A follow-up helical contrast-enhanced thoraco-abdo-pelvic CT scan demonstrated a well-delineated, spontaneously hypodense mass in the left adrenal bed measuring 19 x10 mm (CCxT), exhibiting peripheral calcifications and heterogeneous enhancement post-contrast, while maintaining preserved retroperitoneal fat planes with the left kidney and pancreatic tail. The liver showed a normal height span (8 cm) but marked heterogeneity due to multiple nodular and plaque-like hypodense metastases with peripheral rim enhancement, including measurable target lesions in Segment III (7.6 x7 mm) and Segment VI (10 x 9 mm). Thoracic acquisitions revealed a localized focal area of pulmonary consolidation in the right posterior lung without evidence of parenchymal metastases or pleural effusion. Associated findings included peripheral pancreatic calcifications, whereas the portal vein trunk remained widely patent. Crucially, no pathologic deep lymphadenopathy, organomegaly, or lytic cortical bone lesions were identified on dedicated bone windows, confirming the absence of high-risk metastatic features.

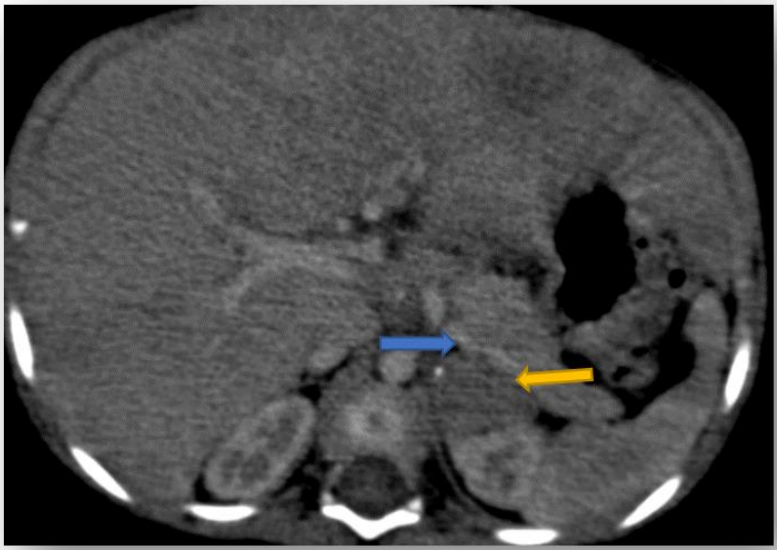

Figure 1: Axial CT image showing well-defined, smooth-contoured mass of the left adrenal fossa, hypodense on unenhanced CT (yellow arrow), exhibiting a few peripheral calcifications (blue arrow)

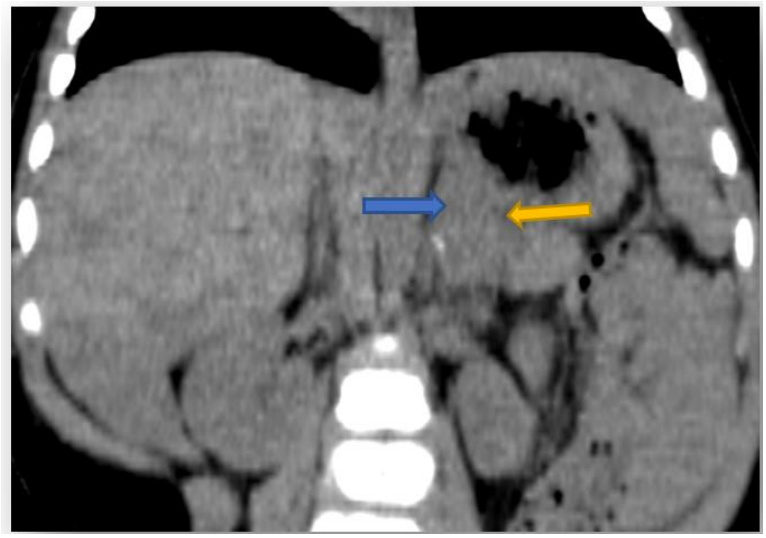

Figure 2: Coronal CT image showing well-defined, smooth-contoured mass of the left adrenal fossa, hypodense on unenhanced CT (yellow arrow), exhibiting a few peripheral calcifications (blue arrow)

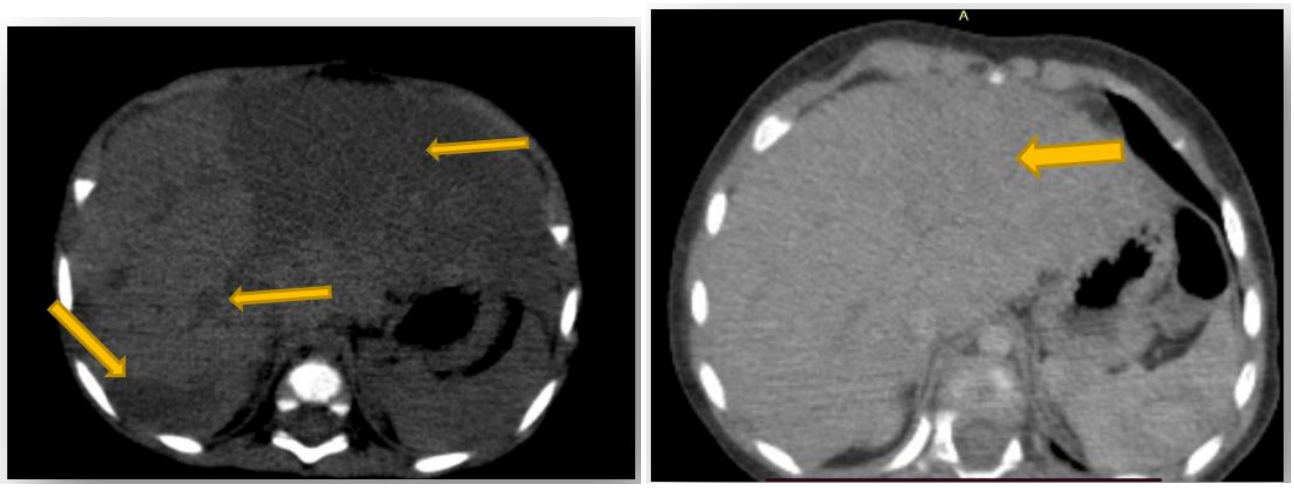

Figure 3: Axial CT images showing heterogeneous liver containing multiple nodular and geographic lesions (yellow arrow), hypodense on unenhanced CT, demonstrating peripheral enhancement following contrast injection

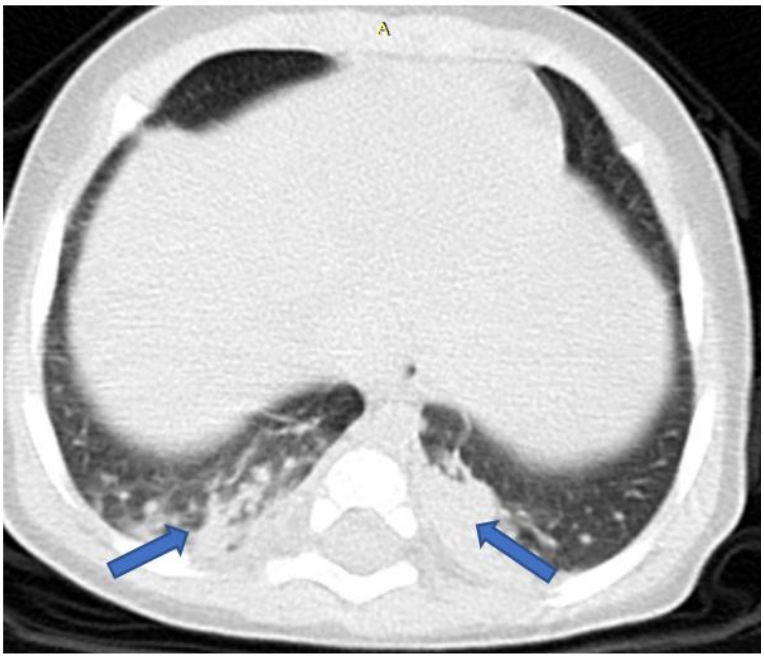

Figure 4: Axial CT images showing bilateral foci of consolidation in the posterior region (blue arrow).

DISCUSSION

Pepper syndrome represents a distinct biological sub-entity within the spectrum of neuroblastic tumors [1]. Unlike typical high-risk stage 4 neuroblastoma—which exhibits aggressive disseminated metastatic disease to cortical bones, bone marrow, distant lymph nodes, and liver in older children—stage 4S (and INRG stage MS) occurs almost exclusively in infants under 12 to 18 months [3, 11]. The underlying biology of stage 4S neuroblastoma is characterized by non-amplified *MYCN* genes, favorable hyperdiploid genomic patterns, and high expression of the TrkA nerve growth factor receptor [5, 9]. These biological hallmarks predispose the metastatic neuroblasts within the hepatic parenchyma to undergo spontaneous differentiation into mature ganglioneuromas or complete cellular apoptosis, leading to spontaneous disease clearance without aggressive cytotoxic intervention [10, 18].

However, despite this favorable intrinsic biology, Pepper syndrome presents a clinical paradox: the sheer physical volume of hepatic tumor burden can precipitate mechanical failure [6]. Rapid expansion of liver metastases leads to severe hepatomegaly, resulting in increased intra-abdominal pressure [16]. This can compromise diaphragmatic excursion, leading to restrictive respiratory insufficiency, compression of the inferior vena cava and renal veins, and abdominal compartment syndrome [6, 16]. Hence, imaging modalities must balance verifying favorable biological distribution against detecting dangerous organ compression.

Multi-Detector Computed Tomography (MDCT) is a cornerstone cross-sectional modality for evaluating abdominal neuroblastoma [7, 8]. On pre-contrast CT, neuroblastomas typically present as lobulated, unencapsulated, hypodense soft-tissue masses originating in the retroperitoneum or adrenal gland [12, 13]. A classical pathognomonic feature is the presence of calcifications—seen in 80% to 90% of neuroblastomas on CT—which can appear as coarse, amorphous, fine punctate, or peripheral ring-like calcifications [14]. In our patient, the left adrenal mass demonstrated characteristic peripheral calcifications and heterogeneous enhancement following iodinated contrast administration, corroborating the diagnosis of primary neuroblastoma.

Post-contrast MDCT acquisitions are equally essential for mapping the unique metastatic liver dissemination in Pepper syndrome [15]. Hepatic metastases in Pepper syndrome can present as either diffuse multinodular low-attenuation lesions or continuous coalescent tumor sheets that replace normal liver parenchyma [8, 15]. On contrast-enhanced images, these lesions typically display peripheral rim enhancement with central hypodensity, representing hypo-vascular tumor necrosis or myxoid stromal changes [14, 15]. In our case, the CT demonstrated a multi-focal nodular pattern with peripheral enhancement, allowing clear identification and baseline quantification of Target 1 (7.6 × 7 mm in segment III) and Target 2 (10 × 9 mm in segment VI). Furthermore, CT revealed peripheral calcifications within the pancreas, an uncommon finding that may represent dystrophic calcification or neuroblastic infiltration, warranting close radiological monitoring.

Distinguishing Stage 4S (INRG Stage MS) from standard Stage 4 neuroblastoma is one of the most vital tasks in pediatric oncology, as it shifts treatment from conservative monitoring/low-dose therapy to intensive

multi-agent chemotherapy, surgical resection, and autologous stem cell transplantation [3, 11]. The presence of distant cortical bone metastases reclassifies an infant from Stage 4S to Stage 4 [11, 12]. High-resolution CT bone window settings provide sensitive evaluation of the cortical skeleton, identifying subtle lytic, sclerotic, or periosteal reactions [8, 12]. In our patient, the complete absence of lytic cortical lesions on CT bone windows supported the Stage 4S diagnosis]. Additionally, contrast-enhanced CT confirmed portal vein patency and the absence of deep pathological adenopathy, further reinforcing a favorable stage profile.

CONCLUSION

Computed Tomography is essential for the diagnosis, initial staging, and longitudinal surveillance of Pepper syndrome. By offering accurate primary tumor characterization, baseline target lesion dimensions, and identification of intercurrent pulmonary or vascular complications, MDCT guides optimal therapeutic management in infants with stage 4S disease.

REFERENCES

1. Maris JM. Recent advances in neuroblastoma. *N Engl J Med*. 2010;362(23):2202-2211.
2. D'Angio GJ, Evans AE, Koop CE. Special pattern of widespread neuroblastoma with a favorable prognosis. *Lancet*. 1971;297(7708):1046-1049.
3. Brodeur GM, *et al.,*Revisions of the international criteria for neuroblastoma diagnosis, staging, and response to treatment. *J Clin Oncol*. 1993;11(8):1466-1477.
4. Pepper W. A study of congenital sarcoma of the liver and suprarenal. *Am J Med Sci*. 1901;121(3):284-299.
5. Taggart DR, *et al.,*Outcomes of infants with stage 4S neuroblastoma: A report from the Children's Oncology Group. *Eur J Cancer*. 2011;47(7):1090-1098.
6. Guglielmi R, *et al.,*Management and outcome of Pepper syndrome in infants: A single-center experience. *Pediatr Blood Cancer*. 2020;67(4):e28150.
7. Brisse HJ, *et al.,*Guidelines for imaging and staging of neuroblastic tumors: consensus report from the International Neuroblastoma Risk Group Project. *Radiology*. 2011;261(1):243-257.
8. Lonergan GJ, *et al.,*Neuroblastoma: clinical-radiologic review. *RadioGraphics*. 2002;22(4):911-934.
9. Schleiermacher G, *et al.,*Clinical relevance of localized stage 4S neuroblastoma. *Br J Cancer*. 2004;91(11):1950-1958.
10. Nickerson HJ, *et al.,*Favorable biology and outcome of stage 4S neuroblastoma. *J Clin Oncol*. 2000;18(3):477-486.
11. Monclair T, *et al.,*The International Neuroblastoma Risk Group (INRG) staging system. *J Clin Oncol*. 2009;27(2):298-303.
12. Siegel MJ. Pediatric body CT. 2nd ed. Philadelphia: Lippincott Williams & Wilkins; 2008.
13. Kazerooni EA, *et al.,*Neuroblastoma: imaging evaluation and staging. *AJR Am J Roentgenol*. 1990;155(6):1265-1270.
14. Rha SE, *et al.,*Neurogenic tumors in the abdomen: imaging findings and pathologic correlation. *RadioGraphics*. 2003;23(1):29-43.
15. Dumba M, *et al.,*Neuroblastoma and nephroblastoma: a practical guide to imaging in pediatric oncology. *Cancer Imaging*. 2015;15(1):5.
16. Hsu E, *et al.,*Abdominal compartment syndrome secondary to massive hepatomegaly in stage 4S neuroblastoma. *J Pediatr Surg Case Rep*. 2018;35:12-15.
17. Eisenhauer EA, *et al.,*New response evaluation criteria in solid tumours: revised RECIST guideline (v1.1). *Eur J Cancer*. 2009;45(2):228-247.
18. Hero B, *et al.,*Localized infant neuroblastomas often show spontaneous regression: results of NB95-S. *J Clin Oncol*. 2008;26(9):1504-1510.